Classification of Tumours of the Central Nervous system based on WHO classification 2021 and ICD-O-3.2 (to be used for new registrations from incidence year 2022)

WHO classification of tumours of central nervous system	Classification ICD-O-3.2	Topography (most frequent)	Comments (all additional information: biomarkers,...)
Gliomas, glioneuronal tumours and neuronal tumours			
Adult-type diffuse gliomas			
Astrocytoma, IDH-mutant		(C71._)	
Astrocytoma, IDH-mutant, grade 2	9400/3	(C71._)	
Astrocytoma, NOS	*9400/3*	*(C71._)*	*NOS*
Astrocytoma, IDH-mutant, grade 3	9401/3	(C71._)	
Astrocytoma, IDH-mutant, grade 4	9445/3	(C71._)	
Gemistocytic astrocytoma, IDH-mutant, NOS	*9411/3*	*(C71._)*	
Oligodendroglioma, IDH-mutant and 1p/19q-codeleted		(C71._)	
Oligodendroglioma, IDH-mutant and 1p/19q-codeleted, grade 2	9450/3	(C71._)	
Oligodendroglioma, NOS	*9450/3*	*(C71._)*	*NOS*
Oligodendroglioma, IDH-mutant and 1p/19q-codeleted, grade 3	9451/3	(C71._)	
Glioblastoma, IDH-wildtype	9440/3	(C71._)	
Giant cell glioblastoma	9441/3	(C71._)	
Gliofibroma	*9442/3*	*(C71._)*	
Gliosarcoma	9442/3	(C71._)	
(Anaplastic) oligoastrocytoma, NOS or NEC	*9382/3*	*(C71._)*	
Paediatric-type diffuse low-grade gliomas			
Diffuse astrocytoma, *MYB* or *MYBL1* -altered*	9421/1	(C71._)	"*MYB* -altered" or "*MYBL1* -altered"
Angiocentric glioma	9431/1	(C71._)	
Polymorphous low-grade neuroepithelial tumour of the young (PLNTY)*	9413/0	(C71._)	
Diffuse low-grade glioma, MAPK pathway-altered*	9421/1	(C71._)	"MAPK-pathway altered" (*BRAF* or *FGFR1*)
Paediatric-type diffuse high-grade gliomas			
Diffuse midline glioma, H3 K27-altered*	9385/3	(C71._)	"Diffuse midline glioma, H3 K27-altered"
Diffuse hemispheric glioma, H3 G34-mutant*	9385/3	(C71._)	"Diffuse hemispheric glioma, H3 G34-mutant"
Diffuse paediatric-type high-grade glioma, H3-wildtype and IDH-wildtype	9385/3	(C71._)	"Diffuse paediatric-type high-grade glioma, H3-wildtype and *IDH* -wildtype"
Infant-type hemispheric glioma*	9385/3	(C71._)	"Infant-type hemispheric glioma"
Circumscribed astrocytic gliomas			
Pilocytic astrocytoma	9421/1	(C71._; C72._)	Pilocytic astrocytoma (*KIAA1549::BRAF* fusion)
Pilomyxoid astrocytoma	9425/3	(C71._)	
High-grade astrocytoma with piloid features (HGAP)	9421/3[5]	(C71._)	
Pleomorphic xanthoastrocytoma (PXA)	9424/3	(C71._)	
Subependymal giant cell astrocytoma (SEGCA)	9384/1	(C71._)	
Chordoid glioma	9444/1	(C71._)	
Astroblastoma, MN1-altered*	9430/3	(C71._)	
Glioma, NOS / NEC	*9380/3*	*(C71._)*	*To be justified with all genetic/molecular aberrations, NEC/NOS*
Gliomatosis cerebri, NOS	*9381/3*	*(C71._)*	
Glioneuronal and neuronal tumours			
Ganglioglioma	9505/1	(C70._; C71._; C72._)	
Anaplastic ganglioglioma	9505/3	(C70._; C71._; C72._)	
Gangliocytoma	9492/0	(C70._; C71._; C72._)	
Desmoplastic infantile ganglioglioma	9412/1	(C71._)	"Desmoplastic infantile ganglioglioma"
Desmoplastic infantile astrocytoma	9412/1	(C71._)	"Desmoplastic infantile astrocytoma "
Dysembryoplastic neuroepithelial tumour (DNET)	9413/0	(C71._)	
Diffuse glioneuronal tumour with oligodendroglioma-like features and nuclear clusters (DGONC) (provisional entity)	Not applicable		
Papillary glioneuronal tumour (PGNT)	9509/1	(C71._; C75.3)	"Papillary glioneuronal tumour"
Rosette-forming glioneuronal tumour (RGNT)	9509/1	(C71._)	"Rosette-forming glioneuronal tumour"
Myxoid glioneuronal tumour*	9509/1	(C71._)	"Myxoid glioneuronal tumour"
Diffuse leptomeningeal glioneuronal tumour	9509/3[5]	(C70._; C71;_; C72._)	
Multinodular and vacuolating neuronal tumour	9509/0[5]	(C71._)	
Dysplastic cerebellar gangliocytoma (Lhermitte-Duclos disease)	9493/0	(C71.6)	
Central neurocytoma	9506/1	(C71._; C72._)	"Central neurocytoma"
Extraventricular neurocytoma	9506/1	(C71._)	"Extraventricular neurocytoma"
Cerebellar liponeurocytoma	9506/1	(C71.6)	"Cerebellar liponeurocytoma"
Ependymal tumours			
Supratentorial ependymoma, *ZFTA* fusion-positive*	9396/3	(C71._)	"Supratentorial ependymoma, *ZFTA* fusion-positive"
Supratentorial ependymoma, *YAP1* fusion-positive*	9396/3	(C71.5)	"Supratentorial ependymoma, *YAP1* fusion-positive"
Supratentorial ependymoma, NOS*	9391/3	(C71._)	"Supratentorial"
Posterior fossa group A (PFA) ependymoma*	9396/3	(C71.7)	"PFA"
Posterior fossa group B (PFB) ependymoma*	9396/3	(C71.7)	"PFB"
Posterior ependymoma, NOS*	9391/3	(C71.7)	"Posterior"
Subependymoma	9383/1	(C71.7)	
Spinal ependymoma, MYCN-amplified*	9396/3	(C72._)	"Spinal ependymoma, *MYCN* -amplified"
Spinal ependymoma, NOS*	9391/3	(C72._)	"Spinal"
Myxopapillary ependymoma	9394/1	(C72._)	
Sellar ependymoma	9391/1	(C75.1)	
Choroid plexus tumours			
Choroid plexus papilloma	9390/0	(C71.5; C71.7)	
Atypical choroid plexus papilloma	9390/1	(C71.5; C71.7)	
Choroid plexus carcinoma	9390/3	(C71.5; C71.7)	
Embryonal tumours			
Medulloblastomas, molecularly defined			
Medulloblastoma, WNT-activated	9475/3	(C71.6)	
Medulloblastoma, SHH-activated and *TP53* -wildtype	9471/3	(C71.6)	"SHH-activated and *TP53* -wildtype"
Medulloblastoma, SHH-activated and *TP53* -mutant	9476/3	(C71.6)	
Medulloblastoma, non-WNT/non-SHH	9477/3	(C71.6)	
Medulloblastomas, histologically defined			
Desmoplastic nodular medulloblastoma	9471/3	(C71.6)	"Desmoplastic nodular"
Medulloblastoma with extensive nodularity	9471/3	(C71.6)	"Extensive nodularity"
Large cell medulloblastoma	9474/3	(C71.6)	"Large cell"
Anaplastic medulloblastoma	9474/3	(C71.6)	"Anaplastic"
Medulloblastoma, histologically defined	9470/3	(C71.6)	
Medullomyoblastoma	9472/3	(C71._)	
Other CNS embryonal tumours			
Atypical teratoid/rhabdoid tumour (=ATRT)	9508/3	(C71._)	
Cribriform neuroepithelial tumour (CRINET) (provisional entity)	Not applicable		
Embryonal tumour with multilayered rosettes (ETMR)	9478/3	(C71._)	
CNS neuroblastoma, *FOXR2* -activated*	9500/3	(C71._;C72._)	"CNS neuroblastoma, *FOXR2* -activated"
Ganglioneuroblastoma	9490/3	(C71._;C72._)	
CNS tumour with *BCOR* internal tandem duplication*	9500/3	(C71._)	"CNS tumour with *BCOR* internal tandem duplication"
CNS embryonal tumour, NEC/*NOS*	9473/3	(C71._;C72._)	
Medulloepithelioma	9501/3	(C71._;C72._)	
Pineal tumours			
Pineocytoma	9361/1	(C75.3)	
Pineal parenchymal tumour of intermediate differentiation (PPTID)	9362/3	(C75.3)	"Pineal parenchymal tumour of intermediate differentiation"
Pineoblastoma	9362/3	(C75.3)	"Pineoblastoma"
Papillary tumour of the pineal region	9395/3	(C75.3)	
Desmoplastic myxoid tumour of the pineal region, *SMARCB1* -mutant (provisional	Not applicable		
Cranial and paraspinal nerve tumours			
Schwannoma	9560/0	(C72._)	
Neurofibroma	9540/0	(C72._)	
Plexiform neurofibroma	9550/0	(C72._)	
Perineurioma	9571/0	(C72._)	
Hybrid nerve sheath tumour	9563/0	(C72._)	
Malignant melanotic nerve sheath tumour	9540/3	(C72._)	"Melanotic"
Malignant peripheral nerve sheath tumour (MPNST)	9540/3	(C72._)	"Peripheral"
Epitheliod MPNST	9540/3	(C72._)	'Epitheliod MPNST''
Malignant perineurioma	9571/3	(C72._)	
Cauda equina neuroendocrine tumour (previously paraganglioma)	8693/3	(C72.1)	
Meningioma			
Meningioma	9530/0	(C70._)	*Information about atypical/anaplastic must be registrated using the CNS WHO grade*
Meningiothelial meningioma	9531/0	(C70._)	
Fibrous meningioma	9532/0	(C70._)	
Transitional meningioma	9537/0	(C70._)	
Psammomatous meningioma	9533/0	(C70._)	
Angiomatous meningioma	9534/0	(C70._)	
Hemangioblastic meningioma	9535/0	(C70._)	
Chordoid or clear cell meningioma	9538/1	(C70._)	
Papillary or rhabdoid meningioma	9538/3	(C70._)	

Atypical meningioma	*9539/1*	*(C70._)*	*By default if not possible to subclassify by morphology (Information about atypical/anaplastic must be registrated using the CNS WHO grade)*
Anaplastic malignant meningioma	*9530/3*	*(C70._)*	
Mesenchymal, non-meningothelial tumours involving the CNS			
Fibroblastic and myofibroblastic tumours			
Solitary fibrous tumour (SFT; previously termed hemangiopericytoma)	8815/1	(C70._; C71._; C72._)	
Vascular tumours			
Cavernous haemangioma	9121/0	(C70._; C71._; C72._)	
Capillary haemangioma	9131/0	(C70._; C71._; C72._)	
Arteriovenous malformation (=AVM)	9123/0	(C70._; C71._; C72._)	
Haemangioblastoma	9161/1	(C70._; C71._; C72._)	
Skeletal muscle tumours			
Embryonal rhabdomyosarcoma	8910/3	(C70._; C71._; C72._)	
Alveolar rhabdomyosarcoma	8920/3	(C70._; C71._; C72._)	
Rhabdomyosarcoma, pleomorphic-type	8901/3	(C70._; C71._; C72._)	
Spindle cell rhabdomyosarcoma	8912/3	(C70._; C71._; C72._)	
Tumours of uncertain differentiation			
Intracranial mesenchymal tumour, FET::CREB fusion-positive (provisional entity)	Not applicable		
CIC -rearranged sarcoma	8803/3	(C70._; C71._; C72._)	9367/3[$] (Code does not yet exist in ICD-O-3.2)
Primary intracranial sarcoma, *DICER1* -mutant*	9480/3	(C70._; C71._; C72._)	
Ewing sarcoma	9364/3	(C70._; C71._; C72._)	
Chondrogenic tumours			
Mesenchymal chondrosarcoma	9240/3	(C70._; C71._; C72._)	
Chondrosarcoma	9220/3	(C70._; C71._; C72._)	
Dedifferentiated chondrosarcoma	9243/3	(C70._; C71._; C72._)	
Notochordal tumours			
Chordoma	9370/3	(C40._;C41._)	
Melanocytic tumours			
Diffuse meningeal melanocytic neoplasms			
Meningeal melanocytosis	8728/0	(C70._)	
Meningeal melanomatosis	8728/3	(C70._)	
Circumscribed meningeal melanocytic neoplasms			
Meningeal melanocytoma	8728/1	(C70._)	
Meningeal melanomatosis	8720/3	(C70._)	
Melanotic neuroectodermal tumor	9363/0	(C70._)	
Haematolymphoid tumours involving the CNS			
CNS lymphomas			
Primary diffuse large B-cell lymphoma of the CNS	9680/3	(C70._; C71._; C72._)	
Immunodeficiency-associated CNS lymphomas		(C70._; C71._; C72._)	Classify according to the corresponding lymphoma subtype and in comment the corresponding immunodeficiency 'PTLD' or 'HIV' or 'PID'**, ...
Lymphomatoid granulomatosis	9766/1	(C70._; C71._; C72._)	Indicate the grade
Lymphomatoid granulomatosis, grade 1	9766/1	(C70._; C71._; C72._)	"grade 1"
Lymphomatoid granulomatosis, grade 2	9766/1	(C70._; C71._; C72._)	"grade 2"
Lymphomatoid granulomatosis, grade 3	9766/3	(C70._; C71._; C72._)	
Intravascular large B-cell lymphoma	9712/3	(C70._; C71._; C72._)	
Miscellaneous rare lymphomas in the CNS			
MALT lymphoma of the dura	9699/3	(C70._; C71._; C72._)	
Lymphoplasmacytic lymphoma	9671/3	(C70._; C71._; C72._)	
Follicular lymphoma	9690/3	(C70._; C71._; C72._)	
Anaplastic large cell lymphoma ALK+	9714/3	(C70._; C71._; C72._)	
Anaplastic large cell lymphoma ALK-	9715/3	(C70._; C71._; C72._)	
Peripheral T-cell lymphoma (PTCL)	9702/3	(C70._; C71._; C72._)	
NK/T-cell lymphoma (nasal type), with primary manifestation in the CNS	9719/3	(C70._; C71._; C72._)	
Histiocytic tumours			
Erdheim-Chester disease	9749/3	(C70._; C71._; C72._)	"Erdheim-Chester disease"
Rosai-Dorfman disease*	9749/3	(C70._; C71._; C72._)	"Rosai-Dorfman disease"
Juvenile xanthogranuloma*	9749/1	(C70._; C71._; C72._)	
Langerhans cell histiocytosis	9751/1	(C70._; C71._; C72._)	
Histiocytic sarcoma	9755/3	(C70._; C71._; C72._)	
Germ cell tumours			
Mature teratoma	9080/0	(C70._; C71._; C72._)	
Immature teratoma	9080/3	(C70._; C71._; C72._)	
Teratoma, NOS	*9080/1*	*(C70._; C71._; C72._)*	
Teratoma with somatic-type malignancy	9084/3	(C70._; C71._; C72._)	
Germinoma	9064/3	(C70._; C71._; C72._)	
Embryonal carcinoma	9070/3	(C70._; C71._; C72._)	
Yolk sac tumour	9071/3	(C70._; C71._; C72._)	
Choriocarcinoma	9100/3	(C70._; C71._; C72._)	
Mixed germ cell tumour	9085/3	(C70._; C71._; C72._)	
Tumours of the sellar region			
Craniopharyngioma			
Adamantinomatous craniopharyngioma	9351/1	(C75.2)	
Papillary craniopharyngioma	9352/1	(C75.2)	
Craniopharyngioma, NOS	*9350/1*	*(C75.2)*	
Pituicytoma, granular cell tumour of the sellar region, and spindle cell oncocytoma			
Pituicytoma	9432/1	(C75.1)	
Granular cell tumour of the sellar region	9582/0	(C75.1)	
Spindle cell oncocytoma	8290/0	(C75.1)	
Pituitary adenoma/pituitary neuroendocrine tumour (PitNET)*	8272/3	(C75.1)	
Pituitary blastoma	8273/3	(C75.1)	
Unspecified tumours			
Tumour cells, benign	*8000/0*	*(C70._;C71._;C72._)*	
Unclassified tumour, borderline malignancy	*8000/1*	*(C70._;C71._;C72._)*	
Neoplasm, malignant	*8000/3*	*(C70._;C71._;C72._)*	

In grey: codes and / or names of entities which should be avoided in favour of a more specific code

Altered genes are italicized while gene families and proteins are not

*These entities have undergone a change in terminology of a previous code

[$]Codes were approved by the IARC/WHO Committee for ICD-O at its meeting in May 2021

** : PTLD : Post-Transplant Lymphoproliferative disorder; PID : Primary ImmunoDeficiency

These following types and codes must not be used anymore :

Fibrillary astrocytoma	*9420/3*	*(C71._)*	*Mutation status (IDH1/2, 1p/19q codel) takes priority over morphology subtyping for astrocytomas*
Protoplasmic astrocytoma	*9410/3*	*(C71._)*	*Mutation status (IDH1/2, 1p/19q codel) takes priority over morphology subtyping for astrocytomas*
Papillary ependymoma	*9393/3*	*(C71._)*	*Should not be confused with myxopapillary ependymoma. Topography and molecular status take priority over morphology subtyping for ependymomas*
Anaplastic ependymoma	*9392/3*	*(C71._)*	*Replaced by registration of the anaplastic using the CNS WHO grade 3 with the corresponding morphology type and code of ependymoma*
Melanotic schwannoma	*9560/1*	*(C70._)*	*--> 9540/3 : Malignant melanotic nerve sheath tumour*